OUTCOME AND ASSESSMENT FOLLOWING MODIFIED FONTAN REPAIR FOR HYPOPLASTIC LEFT HEART SYNDROME

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We reviewed the clinical outcome and hemodynamic assessment of 74 consecutive pts (age range 5 months - 6 years, median =19 months) who underwent modified Fontan procedure from 8/85-7/89 as reparative surgery for hypoplastic left heart syndrome (HLHS). Additional procedures (n=38) performed in 32 pts *prior* to modified Fontan procedure included shunt revision (n=17), balloon angioplasty of the arch (n=8), atrial septectomy or septostomy (n=6), arch reconstruction (n=4), and pulmonary artery angioplasty (n=3). The types of Fontan included: inferior vena cava baffle within the right atrium to the superior vena caval-pulmonary artery anastomosis, with pulmonary artery augmentation (n=64), or baffle of pulmonary venous return to the tricuspid valve (n=10).

Actuarial survival was 75% (1 month) and 60% (12 months). The median length of hospital stay was 23 days (range 10-112 days). Of the 44 long-term survivors, 18 pts have returned for elective postoperative cardiac catheterization (age mean=34 months, at mean of 16 months following Fontan).

The *hemodynamic findings* were:

	Mean	SD		Mean	SD
Hgb (g/dl)	14.0	(1.0)	SVR (U·m^2)	24.7	(5.6)
SAO_{O2} (%)	94	(3)	RAP (mmHg)	11	(3)
CI (L/min/m2)	2.7	(0.5)	PAP (mmHg)	11	(3)
PVR (U·m^2)	1.8	(0.6)	PAWP(mmHg)	6	(3)

The *angiographic findings* included: mild tricuspid valve regurgitation (n=2), mild native pulmonary valve insufficiency (n=1), and small right to left shunts (systemic venous atrium to pulmonary venous atrium)(n=6). In *all* pts, the pulmonary arteries appeared widely patent and the reconstructed aortic arches appeared unobstructed.

Conclusion: In the subgroup of patients with HLHS who have been evaluated electively by cardiac catheterization following modified Fontan, the utilization of the right ventricle as the systemic ventricle yields excellent intermediate hemodynamic results.

THE USE AND MODIFICATION OF THE GLENN SHUNT

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Forty-eight consecutive pts who underwent Glenn Shunts (SVC-PA Connection) and available for long-term follow-up were reviewed. They included 26 males (54%) and 22 females (46%) whose ages ranged from 7 mos to 46 yrs, mean 7.3 yrs. Diagnoses were single ventricle 28 (58%), tricuspid atresia 12 (25%), transposition 6 (10%), pulmonary atresia 2 (4%) and Ebsteins 1 (2%). 94% pts had prior surgery including BT Shunts 54%, PA Banding 23%, Waterston 6%, Potts 2%, Baffles 2%. Indications included cyanosis in pts 1) whose PA Pressure mean (PAP) was 20-25mm Hg; or 2) whose associated anatomy precluded early Fontan Repair; or 3) whose ventricular ejection fraction by MUGA Scan was ≤ 40%. Pre-operatively, PAP mean was 18.2mm Hg; mean arterial saturation was 79% and mean hematocrit (hcrit) 53.4%. Post-operatively, mean PAP was 16.5mm Hg (NS), arterial saturation 91.6% ($P<.0001$) and hcrit 40% ($P<.0001$). 21 (44%) pts received classic (PA divided) while 27 (56%) received modified (PA undivided) Glenn Shunts. 29 operations (60%) were performed on cardiopulmonary bypass with associated intra-cardiac procedures while 19 (40%) were performed as closed procedures using a temporary innominate vein to atrial shunt.

There was one hospital mortality (2%). There was, however, a significant incidence of post-operative effusions, 17 pts (35%). The mean pre-operative PAP for this group was significantly higher as compared to the group who did not have effusion 19.1 vs 13mm Hg ($P<.05$). In addition, the previous practice of dissecting the SVC-tracheal lymphatics area has been discontinued because of a possible association. To date, 7 pts have undergone subsequent Fontan without mortality or complication.

In summary, the Glenn Shunt 1) can safely be performed in wide range of cyanotic pts where complex anatomy precludes Fontan; 2) provides excellent palliation by significant improvement in post-operative saturation and hcrit; 3) pts previously unacceptable may be safely "bridged" to Fontan completion; and 4) modifications including temporary shunt facilitated closed repair and avoidance of paratracheal dissection during mobilization of SVC may help lessen the incidence of post-operative pleural effusion particularly in pts with high PAPs.

RELIEF OF AORTIC OUTFLOW OBSTRUCTION IN INFANCY: USE OF THE PULMONARY ARTERY TO AORTIC ANASTOMOSIS

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Surgical approaches to aortic outflow obstruction in various forms of complex congenital heart disease have included conduit placement, PA banding, and subaortic resection with a reported survival of 50% in all age groups. We investigated the use of PA to ascending aorta anastomosis (PAA) as relief of aortic outflow obstruction in *infancy* and determined if the anatomic sub-type affected early survival.

Between December 1985 and August 1989, 47 pts underwent PAA at a median age of 10 days (range 2-184 days). Pts were divided into 3 anatomic subgroups: Group I (n=17) had aortic stenosis/atresia with ventricular septal defect (VSD), 11 of these had normal mitral valve and LV, 6 had mitral stenosis; Group II (n=24) had univentricular hearts of LV morphology, 16 were L loop and 8 D loop; Group III (n=6) had double outlet RV with subaortic stenosis, 2 of these had normal mitral valve and LV. Surgery consisted of PAA with pulmonary homograft augmentation of the arch, atrial septectomy and either a 4mm central shunt (37) or a modified Blalock-Taussig shunt (7). Three infants underwent initial biventricular repair (BVR) consisting of PAA with simultaneous closure of the VSD to the pulmonary valve and placement of a RV to PA conduit.

There were 32 survivors with a post-operative follow up period of 3 to 30 months. Early survival (30 days) consisted of 14 in Group I (82%), 18 in Group II (75%), and 5 in Group III (83%) with no significant difference between the groups. The overall actuarial survival at 1 year was 68%. Of the 13 infants in Groups I and III who had normal mitral valve and LV, 2 died after initial BVR and 2 died after PAA; 1 survived initial BVR, 3 have undergone PAA with subsequent successful BVR, and 5 are awaiting further corrective surgery. Of the remainder, 8 pts have gone on to modified Fontan procedure, and 3 to a modified superior caval to pulmonary anastomosis in anticipation of a final procedure.

CONCLUSIONS: PAA can be successfully employed in infancy with equal early survival in a variety of anatomic subgroups with aortic outflow obstruction, and allows for flexibility in the choice of subsequent corrective surgery.

IMPORTANT ROLES OF TRANSESOPHAGEAL COLOR DOPPLER FLOW MAPPING STUDIES(TEE) IN INFANTS WITH CONGENITAL HEART DISEASE.

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We performed color Doppler TEE studies in 31 infants aged 1 day to 5 mos (2.21 to 17.8 kgs) for intraoperative evaluation of repairs of congenital heart disease (CHD)(n=27), guidance of cardiac catheterization procedures (n=2) or elucidation of important diagostic features in ventilated infants in the ICU (n=2). In our infants the esophagus could easily be entered with a 6.8mm 5MHz phased array (Aloka/Corometrics) probe except for 1 infant with truncus arteriosus and an AO arch abnormality. The most important intraoperative roles identified were for guiding repairs of AV septal defects (n=11) including 1 with hypoplastic LV. In 3 AV defect infants, surgery was revised based on TEE observations to repair the mitral valve in 2 and in 1 to create a small ASD to vent the LA. In 9 infants with VSD's, patch integrity was monitored by TEE. In 2, significant VSD patch leaks and in 1 an additional muscular VSD were identified and repaired. In 4 infants with TOF, and 3 patients with complex conduit repairs, patch integrity, RVOT flow and anatomy were detailed. Other surgical, catheterization related and diagnostic roles for TEE included monitoring surgical atrial septectomies, guiding balloon septostomies, guiding ASD occluding device placement for residual shunting after a Fontan procedure, finding PV's and diagnosing SBE caused patch dehisience in a final critically ill infant. TEE was easy to perform and provided easily obtainable, important information with better image quality and orientation than epicardial studies.